

## **A Nightmare for Emergency Department Physicians: Left-Sided Acute Appendicitis. A Case Report**

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**Received:** December 18, 2019; **Published:** January 04, 2020

### **Abstract**

Appendicitis with acute abdominal pain is one of the commonest presentation in an emergency department. Approximately 30 % patients have an atypical clinical presentation. Atypical location of the appendix (e.g. left side of abdomen), is associated with high risk of misdiagnosis. High level of clinical suspicion and awareness of atypical appendicitis can help physicians to avoid pitfalls in the diagnosis and management of this rare variant.

**Keywords:** *Left Sided Appendicitis; Situs Inversus Totalis; Midgut Malrotation*

### **Abbreviations**

LSA: Left Sided Appendicitis; SIT: Situs Inversus Totalis; MM: Midgut Malrotation; US: Ultrasound; CT: Computed Tomography; VR: Volume Rendering; MIP: Maximum Intensity Projection

### **Introduction**

Appendicitis is one of the most common surgical conditions presenting with acute abdominal pain in an Emergency Department [1-3]. Preoperative diagnosis of acute appendicitis primarily depends upon imaging findings with different clinical features and laboratory investigations playing a supporting role [4]. However, imaging diagnosis of appendicitis is not always simple and straightforward and can be quite difficult, particularly, in patients with atypical clinical presentation and atypical anatomical location of the appendix, which is associated with high risk of missed or delayed diagnosis, leading to increased morbidity and mortality [1,3-5].

Left sided appendicitis (LSA), a rare pathology, is an example of atypical clinical presentation and location of the appendix which needs increased cautiousness and awareness among referring emergency physicians, surgeons and interpreting radiologists to avoid pitfalls and delays in its diagnosis and management [1]. Two situations may explain this rare entity: midgut malrotation or situs inversus. Its diagnosis is suspected by clinical presentation and confirmed by radiological examinations. Imaging allows precise positive preoperative diagnosis and helps in guiding its surgical management.

We report a 14 years male with previously unknown bowel malrotation with a left sided acute appendicitis. CT played a key role in making this otherwise elusive diagnosis.

### **Case Presentation**

14 years old boy brought to the emergency department with 10 hours history of left lower abdominal pain. Pain was acute in onset, moderate in severity, colicky in nature and localized in left lumbar region. There was history of an episode of vomiting (food contents).

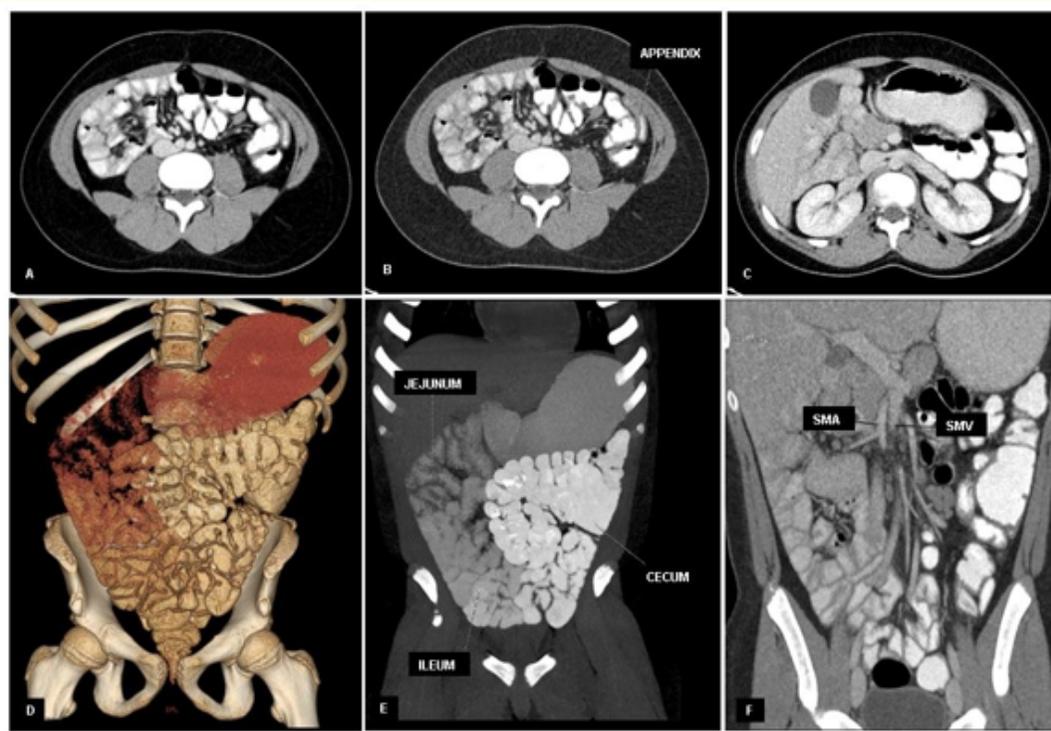
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**Citation:** Ammar Ashraf, *et al.* "A Nightmare for Emergency Department Physicians: Left-Sided Acute Appendicitis. A Case Report". *EC Clinical and Medical Case Reports* 3.2 (2020): 01-05.

There was also history of burning micturition and history of raw camel milk ingestion. No history of fever, rigors or change in bowel habits. No significant past medical history.

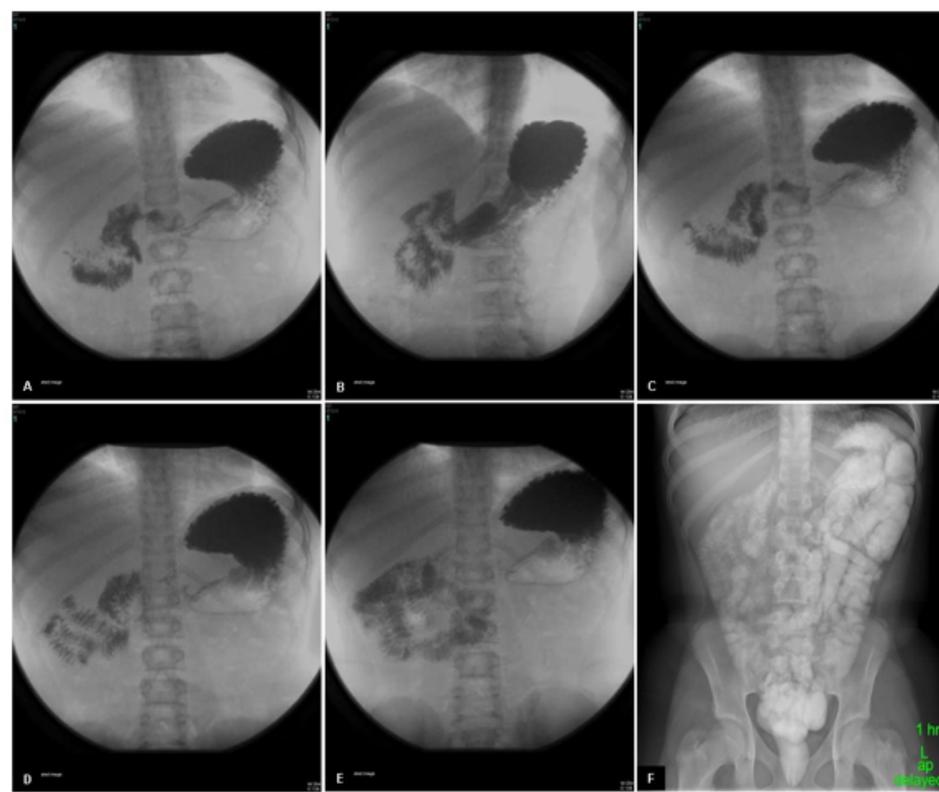
On examination, he was afebrile (36.8°C). His BP was 118/64 mm Hg, heart rate was 84 beats per minute. He had soft lax abdomen with mild tenderness in left lumbar region. Rest of the clinical examination was unremarkable. Laboratory investigations showed leukocytosis; white blood cell (WBC) count was  $20 \times 10^9/L$  ( $4-12 \times 10^9/L$ ) with neutrophil count measuring  $15.58 \times 10^9/L$  ( $1.4-7.7 \times 10^9/L$ ). His c-reactive protein (CRP) was 20.80 mg/dl ( $\leq 1.2$  mg/dl). His chest and abdominal radiographs were within normal limits. Appendix was not visualized on abdominal ultrasound examination; solid abdominal organs including kidneys were normal. No intra-abdominal free fluid/collection was seen.

On call surgical team saw the patient in emergency department and after exclusion of acute surgical causes of abdominal pain, patient was referred to on call internal medicine team. Medical team admitted the patient as a suspected case of urinary tract infection/pyelonephritis, despite normal urine analysis. Next day due to continuous abdominal pain, contrast enhanced CT abdomen (Figure 1) was requested which showed thick walled inflamed appendix measuring 9 mm, located in left lumbar region. No appendicolith, fat stranding, free fluid or collection was seen. There was incidental finding of bowel non-rotation, with small bowel in right abdomen, large bowel in left abdomen, reversal of normal relationship of superior mesenteric vessels and long and broad small bowel mesentery suggesting type Ia malrotation.



**Figure 1:** CT abdomen with oral and IV contrast. Axial images (A, B) show thickened inflamed left sided appendix. Axial (C) and coronal (F) images show reversed SMA and SMV relation. VR and coronal MIP (D, E) images show small bowel loops in the right abdomen and large bowel loops in the left abdomen.

After CT scan, he underwent an uneventful laparoscopic appendectomy. No complications were seen. CT findings of bowel non-rotation with a long and broad small bowel mesentery (type Ia) were noted again during surgery for which no intervention was done as it has no risk for any complications in the future. Postoperative hospital stay was smooth and unremarkable. He was discharged on oral antibiotics and analgesics on 2<sup>nd</sup> postoperative day. Histopathology of the appendix showed acute appendicitis. He was seen in surgical outpatient after one week of surgery with good recovery. Small bowel follow through was done after the surgery which showed duodeno-jejunal flexure on the right side of the midline with all small bowel loops in the right abdomen and colon in the left abdomen (Figure 2).



**Figure 2:** Small bowel follow through showing DJ flexure and small bowel loops in the right abdomen and large bowel loops in the left abdomen (type Ia malrotation).

## Discussion

Appendicitis is one of the commonest surgical causes of lower abdominal pain, particularly around the umbilicus and in right iliac fossa; however, approximately 1/3<sup>rd</sup> cases of appendicitis can have several atypical presentations [1,2]. In such atypical cases, diagnosis of acute appendicitis can be quite problematic and different imaging investigations (plain films, US, CT, bowel studies with contrast) and increased awareness among referring physicians/surgeons and reading radiologists are mandatory for appropriate and timely diagnosis [1].

Pain and tenderness in left abdomen due to appendicitis is an extremely rare condition, which can occur either due to a true left sided appendix seen in midgut malrotation and situs inversus totalis or a wandering cecum and an extremely long right sided appendix crossing the midline [1,6].

In fetus, primitive bowel herniates out of the abdominal cavity and undergoes a 270 degree counter clockwise rotation around the superior mesenteric vessels. Following this rotation, bowel goes back into the abdominal cavity and leads to the next step that is fixation of the colon. After a complete rotation and fixation; duodenojejunal flexure is located at left side of the midline (at the level of first lumbar vertebra), ileocecal junction is in the right iliac fossa and broad base mesenteric root with superior mesenteric artery (and vein) is lying between these two landmarks [5-8]. Any interruption in this rotation and fixation process of the bowel is known as bowel malrotation [1,5,6,8].

Approximately 50% of cases of bowel malrotation are diagnosed during the first week of life while 60% are discovered within the first month of life due to its associated complications like midgut volvulus [1,6]. Rest of the cases may present in childhood or adulthood either as an incidental finding during the investigation of some other abdominal complaints such as appendicitis or due to some associated complications like bowel obstruction due to volvulus [1,9,10].

Previously, bowel malrotation was thought to be a disease of infancy with infrequent cases seen beyond one year of age; however, this concept is changing and recent study on about 200 cases found that 31% patients were infants, 21% were in the age group of 1 to 18 years and the remaining 48% patients were adults [5-8,11].

Malrotation is divided into different types depending on stage of interruption of the embryological process of rotation and fixation of the bowel; for example, defects in early stage of rotation lead to type Ia malrotation or non-rotation where the proximal small bowel lies on the right side of the abdomen and the cecum on the left side of the abdomen [1,12]. Estimated incidence of intestinal non-rotation is 1 in every 500 live births (0.2%; range 0.03 - 0.5%) [1,6,12]. Type Ia is the commonest and the least problematic rotational anomaly because of the broad and long mesenteric root and is most likely diagnosed as an incidental finding during imaging evaluation of some other abdominal pathologies [1,12].

Situs inversus is a rare condition and is of two types: complete (situs inversus totalis-SIT) when both thoracic and abdominal organs are mirrored from their normal orientation and partial (situs inversus partialis) when only one of these cavities is affected <sup>1</sup>. Collin D studied 71000 human appendix samples and mentioned that the incidence of appendicitis associated with SIT is 0.016% [1]. The most extensive study of LSA published in literature, reviewed 64 reports of 95 cases and found that 69.4% patients had SIT while 24.2% patients had midgut malrotation [13].

### Imaging

Imaging investigations play a key role in diagnosis of atypical cases and it is important to know the available options in your set up [5,14]. In cases with atypical anatomical location of appendix, diagnosis of acute appendicitis can be quite challenging and both ultrasound as well as computed tomography can provide vital diagnostic information [5,13]. CT has a sensitivity of 87 - 100%, specificity of 89 - 98%, accuracy of 93 - 98%, positive predictive value of 92 - 98% and negative predictive value of 95 - 100% for acute appendicitis [15]. Graded-compression US has a sensitivity of 85%, specificity of 92%, accuracy of 78 - 96%, positive predictive value of 91 - 94% and negative predictive value of 89 - 97% for acute appendicitis [15].

Inversion of normal relation of superior mesenteric artery and vein is considered to be diagnostic of the disease. Zhou. et al. reported a sensitivity of 100%, specificity of 97.6%, accuracy of 98.6%, positive predictive value of 95.8% and negative predictive value of 100% for ultrasound in determining bowel malrotation [1,16].

### Differential diagnosis

Acute appendicitis is usually not included in the possible differentials of left sided abdominal pain which can be due to a number of pathologies including pyelonephritis, renal or ureteric stone, colonic pathology (constipation, irritable bowel syndrome, inflammatory bowel disease), mesenteric adenitis, Meckel's diverticulitis, epiploic appendicitis or left ovarian/fallopian tube pathology (in females) [1,3,5]. These differentials can be excluded with detailed history and clinical examination, along with laboratory and radiological findings [5].

In our case, interestingly, finding of bowel malrotation was missed in the abdominal ultrasound examination. Referring physician as well as the surgical team was surprised with this rare diagnosis established by CT scan.

### Conclusion

LSA is a rare condition that is not usually entertained in the differential diagnosis of left sided abdominal pain. It is a diagnostic dilemma and often leads to diagnosis and management delays. Thorough medical history, detailed physical examination, laboratory and imaging investigations including US and CT scan and above all, increased experience and awareness among emergency physicians, surgeons and radiologists are mandatory tools for an accurate and timely diagnosis and effective management of such patients.

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**Volume 3 Issue 2 February 2020**

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